Sir,

Sandhu *et al.*[1] showed convincingly that oral rehydration with a glucose-polymer solution (12·5 g/100 ml, giving 730 mmol/l glucose) and 90 mmol/l sodium predisposes to hypernatraemia. In so doing they reproduced the circumstances that contributed to the epidemic of hypernatraemia in the 1950s.[2] At that time commercial oral rehydration solutions were changed to high concentrations of glucose polymers, and probably caused hypernatraemia despite an unexceptional concentration of sodium, 50 mmol/l. The imbalanced ratio of glucose to sodium, and the high concentration of the former, cause osmotic loss of water from the intestines,[2] and increase stool loss (well demonstrated in the study by Rodriguez *et al.*[3] cited also by Sandhu *et al.*).

The authors mis-stated my analysis[2] of the nutritional benefit of oral rehydration. It is not the glucose content that benefits, as they imply, but the improved appetite for *normal* foods brought about by rapid rehydration and electrolyte repletion. Trying to increase caloric density with an expensive poly-glucose is the wrong approach to nutrition as well as to rehydration.

Rapid oral rehydration with the WHO formula, using the monomer glucose, or sucrose, and early refeeding is a well-proved treatment which does not lead to hypernatraemia even in rotavirus infection.[2]

References

1 Sandhu B K, Jones B J M, Brook C G D, Silk D B A. Oral rehydration in acute infantile diarrhoea with a glucose-polymer electrolyte solution. *Arch Dis Child* 1982; **57**: 152–4.

2 Hirschhorn N. The treatment of acute diarrhea in children. An historical and physiological perspective. *Am J Clin Nutr* 1980; **33**: 637–63.

3 Rodriguez J T, Blanco R, Gray I M. Treatment of acute diarrhoea with oral electrolyte solutions (abstract). *Pediatr Res* 1978; **12**: 440.

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Dr Sandhu and co-workers comment:

It is true that palatability of a glucose-polymer electrolyte solution may prove a problem in older children, but the child referred to by Dr Hughes-Davies as refusing received sufficient to allow rehydration. We agree that glucose-polymer may be incompletely absorbed as may be the case with many 'normal' often starch-containing foods, which are reintroduced immediately after rehydration. Despite evidence of malabsorption of glucose-polymer Rodriguez *et al.*[3] found recovery without hypernatraemia the rule in their subjects. However, we should like to emphasise that, contrary to suggestions in the above letters, we did not recommend the formulation used in our study for widespread use but suggested that further studies using a much lower sodium and glucose-polymer content were required. The statement by Dr Hirschhorn that a sodium content of 50 mmol/l is 'unexceptional' ignores the possible contribution of such levels to the development of hypernatraemia.

Intracranial haemorrhage due to vitamin K deficiency associated with alpha-1-antitrypsin deficiency type Pi Z

Sir,

We read with interest the report by Hope *et al.*[1] describing 3 infants with α-1-antitrypsin deficiency who developed a bleeding diathesis during the first month of life. They stressed the scarcity of such reports. We recently treated a 23-day-old infant with α-1-antitrypsin deficiency and phenotype Pi ZZ who, 12 hours after admission for evaluation of cholestasis, developed seizures, coma, bulging fontanelle, and excessive bleeding from puncture sites. A 9% reduction in haematocrit was noticed and lumbar and ventricular taps were grossly haemorrhagic. He was breast fed and had not received vitamin K at birth. The prothrombin index was low but soon increased after vitamin K administration. Shortly after the haemorrhagic episode, hydrocephalus was noted and a ventriculo-peritoneal shunt had to be implanted. At present (age 20 months) he has left hemiplegia and moderate developmental delay. Liver function tests remain only slightly abnormal.

Intracranial haemorrhage has seldom been quoted as an early complication linked to vitamin K deficiency in α-1-antitrypsin deficiency.[2] This case however, stresses the importance of this potential complication and indicates that an urgent determination of prothrombin index should be performed in any infant admitted for evaluation of neonatal cholestasis.

References

1 Hope P L, Hall M A, Millward-Sadler G H, Normand I C S. Alpha-1-antitrypsin deficiency presenting as a bleeding diathesis in the newborn. *Arch Dis Child* 1982; **57**: 68–70.

2 Sharp H L. Alpha-1-antitrypsin deficiency. In: Lebenthal E, ed. *Digestive diseases in children.* New York: Grune & Stratton, 1978: 237–42.

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Alpha-1-antitrypsin deficiency, bleeding diathesis, and intracranial haemorrhage

Sir,

We should like to report another child who had α-1-antitrypsin deficiency with disordered clotting which presented as a presumed intracranial haemorrhage.

This girl was born at term to unrelated healthy parents and the perinatal period was uneventful. Vitamin K was

given routinely at birth and she was fully breast fed, and was thriving and developing normally. At age 6 weeks she developed a blood-stained nasal discharge and 48 hours later became lethargic, reluctant to feed, and was noticed to be jaundiced. Six hours later she collapsed and had a grand mal convulsion.

When examined she was apyrexial, pale, and jaundiced with a firm liver edge palpable 3 cm below the right costal margin; she was unresponsive and hypertonic with a tense fontanelle. There was excessive bleeding from venepuncture sites and cerebrospinal fluid obtained from lumbar puncture was grossly blood stained. The haemoglobin concentration was 9·8 g/dl with a normal blood film; platelet count was normal but prothrombin time was more than 120 seconds.

Vitamin K and fresh frozen plasma were given. Within 6 hours her haemoglobin concentration fell to 6·8 g/dl. Liver function tests were abnormal (bilirubin 132 μmol/l, unconjugated 80 μmol/l, with increased levels of transaminases) but clotting tests were nearly normal (prothrombin time 19 seconds, control 14 seconds). A provisional diagnosis of acute encephalopathy was made and she was treated intensively with endoctracheal intubation, hyperventilation, and high dose phenobarbitone in an attempt to reduce intracranial pressure. Ultrasound examination showed normal ventricles with no evidence of intraventricular haemorrhage but subdural effusions could not be excluded and as the fontanelle was still tense 28 hours after the initial seizure, bilateral subdural taps were done which showed heavily blood-stained cerebrospinal fluid under pressure on the right side.

There was no subsequent evidence of bacterial or viral infections. Six weeks after the acute illness she had begun to smile again but was generally hypertonic. There had been no increase in head circumference and computerised tomography showed widespread cerebral atrophy with bilateral subdural effusions. Plasma protein electrophoresis showed that the α-1 band was greatly reduced. The concentration of α-1 antitrypsin was 0·5 g/l and phenotype homozygous Pi Z (father: 0·9 g/l, MZ; mother: 1·2 g/l, MZ: adult reference interval 1·8–3·0 g/l). A liver biopsy specimen showed extensive fibrosis with PAS-positive diastase-resistant granules in the periportal zones, and electron microscopy also showed the typical appearance of α-1-antitrypsin deficiency.

We think that the bleeding disorder was caused by the α-1-antitrypsin deficiency and as a result she suffered an intracranial haemorrhage with extension into the subarachnoid space. We confirm the conclusions of Hope *et al.*[1] that α-1-antitrypsin deficiency may present as a bleeding diathesis. Early diagnosis and treatment are needed to prevent devastating sequelae.

Reference

[1] Hope P L, Hall M A, Millward-Sadler G H, Normand I C S. Alpha-1-antitrypsin deficiency presenting as a bleeding diathesis in the newborn. *Arch Dis Child* 1982; **57**: 68–70.

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This correspondence is now closed: *Editor.*

New sign of tracheo-oesophageal fistula?

Sir,

The classical presentation of oesophageal atresia with tracheo-oesophageal fistula is the 'bubbly' baby who has difficulty swallowing his or her own saliva and whose respiration becomes noisy.

I offer a new sign of tracheo-oesophageal fistula, tentative because in clinical medicine new clinical signs are often signs rediscovered, and also because it is based on observations in a single baby. He was born by elective caesarean section at 40 weeks' gestation. There were no clinical signs of polyhydramnios and no excess of amniotic fluid noted when the uterus was incised. Resuscitation was routine although the baby became secondarily apnoeic at 5 minutes and had to be aspirated under direct vision, when a small amount of viscid mucus was obtained. One hour later a similar thing happened but the baby was not 'bubbly'.

An attempt to pass a nasogastric tube failed and *x*-ray films showed the curled up tube in the upper third of the oesophagus. Auscultation of the chest was equal on both sides but the striking feature was that on auscultation of the abdomen, especially the upper half, breath sounds were as easily heard as over the chest, although they had a slightly resonant or amphoric quality. On reflection this would not be surprising in the common type of tracheo-oesophageal fistula where the middle portion of the oesophagus is atretic and the lower third moiety communicates with the trachea. In the baby we described this communication was wide and joined with the trachea at its bifurcation.

I should like to know whether other workers have either noticed this sign or have read about it.

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